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ARCHIVES OF PEDIATRICS

May 1959
Vol. 76 No. 5

JUVENILE RHEUMATOID ARTHRITIS IN NEW YORK CITY

161 Cases

ROBERT H. MANHEIMER, M.D.,*
KATRINE R. C. GREENE AND FRANCES KROLL

New York

This paper reports on a study of juvenile rheumatoid arthritis in a large metropolitan area. While the study's primary objective was to analyze the disease's social and economic impact, so much data was necessarily collected on the prevalence, symptoms, course, treatment and residual effects of the disease that it seems useful to make this material available to physicians specializing in children's diseases.

The study was supervised by a physician (the Medical Director, New York State Chapter, Arthritis and Rheumatism Foundation), the field investigation being carried out under his direction by a social worker experienced in child care and research. The investigator was thoroughly briefed on the natural history of the disease, its terminology and the use of medical records. Medical data, obtained by the investigator from hospital records and conferences with physicians, was regularly reviewed by the study director.

Cases were selected for study if they met the following criteria: children under the age of 19, with onset before the age of 13 and involvement of two or more joints, whose illness had lasted for at least three months or who presented x-ray or joint changes clearly indicative of juvenile rheumatoid arthritis.

CASE FINDING

Cases for the study were sought through 85 voluntary and municipal hospitals in New York City, including all the general hos-

*Medical Director, Arthritis and Rheumatism Foundation.

pitals having arthritis, pediatric or orthopedic services, children's hospitals and chronic disease hospitals with pediatric services. Proprietary, Federal and other specialized hospitals were excluded. These 85 hospitals were asked to search their in-patient diagnostic files and the records of their arthritis, pediatric and orthopedic services for cases. The hospital case records were then reviewed to identify those meeting the criteria. The exhaustiveness of the hospital searches varied, depending on filing and indexing methods and the availability of older records and of staff time. Many hospital-referred cases were excluded because they were rheumatic fever patients or over the study's age limit. Hospitals whose records were searched in 1955 were not rechecked in 1957.

The number of cases found per hospital varied widely. One hospital, a leading teaching institution with large pediatric and arthritis services, referred 48 cases which met the study criteria. Three hospitals contributed from 15 to 25 cases each; three referred from five to eight; and thirteen reported from one to three. Sixty-five hospitals referred no cases which met the criteria. Altogether, 161 cases were found, 157 from these institutions and four from other sources.

Among the 161 children, 113 (70%) were New York City residents at the time of the study; 25 lived elsewhere in New York State; 16 lived in New Jersey or Connecticut; and seven in Arizona, California, Florida, Michigan, Pennsylvania and South Carolina.

In April 1957 the special census of New York City reported some 2,115,000 children under the age of 19. These 113 New York City cases therefore indicate a prevalence of 5.34 per 100,000 children under age 19.

While no claim is made that the study identified all the New York City residents who had juvenile rheumatoid arthritis as defined by the above criteria, the small number is significant. It seems unlikely that substantial numbers of children with the symptoms common to this illness, requiring, as they usually do, special diagnostic work-ups and frequently showing residual changes, could have been overlooked during a 16-month search in cooperation with 85 hospitals in the city, especially since all the large hospitals with arthritis and pediatric clinics were included.

As indicated above, medical records from twenty hospitals constituted the basic source of information. This was supplemented, in over half the cases, by personal conferences with the patient's

physician. In addition, the investigator made home visits to 79 cases. Additional data were obtained in a substantial number of cases from the hospital's social service department and, in three, from other social agencies.

TABLE 1—SOURCES OF INFORMATION

Sources	No. of Pts.
Hospital Medical Records	161
Conferences with Physicians	92
Home Visits	79
Hospital Social Service Depts., etc.	53

The data presented in this paper offer only a minimal picture of the children's medical history. This results from the fact that the comprehensiveness of the records (obtained from 20 hospitals) varied and that many different physicians were involved in the examination and recording of the 161 cases. Consequently, it was not always clear that all findings had been recorded.

THE CHILDREN'S MEDICAL HISTORY

Sex and Age: The group included 113 girls and 48 boys. When the data were compiled (1957) their ages ranged from 3 to 20, about half the group being under 13 years of age. Ten children aged 19 or 20 in 1957 were included because they were under 19 when seen in 1955 or 1956. Four children died before the study began or before the home visit could be made and two, who were visited, died before the report was completed.

TABLE 2—AGE OF CHILDREN LIVING AT END OF STUDY

Age	No. of Pts.
0– 4 years	5
5– 8 "	33
9–12 "	39
13–16 "	42
17–20 "	36
Total	155

The Onset of Juvenile Rheumatoid Arthritis: The earliest age at which onset was recorded was a few weeks (see Table 3), 54% of the patients being under five at onset, while onset occurred before the age of 9 in 81% of the children.

TABLE 3—AGE AT ONSET

Age at Onset	No. of Pts.
0–12 months	2
1– 4 years	85
5– 8 "	44
9–12 "	30
Total	161

As shown in Table 4, most of the children had had juvenile rheumatoid arthritis for at least five years at the time the study was made, two having been ill for 17 years and one for a few months only.

TABLE 4—PERIOD SINCE ONSET

Period Since Onset	Children Living at End of Study
0–12 months	1
1– 4 years	38
5– 8 "	63
9–12 "	30
13–16 "	21
17 "	2
Total	155

The nature of the onset was described in 122 medical records (see Table 5). Onset was acute in almost two thirds of these, i.e., it was manifested by sudden and persistent swelling, pain or limitation of motion, usually accompanied by fever and other systemic symptoms. In slightly over one third of the recorded cases, onset was gradual, the records reporting fleeting, intermittent swelling and pain.

TABLE 5—NATURE OF ONSET

Recorded		122
Acute	78	
Gradual	44	
Not Recorded		39
Total		161

Areas of Involvement: In most of the case records, the disease was described as involving most of the joints of the body, the effects being specifically limited to the upper or lower extremities alone in 16 patients.

TABLE 6—AREAS INVOLVED

Areas Involved	No. of Pts.
Extremities	117
" plus Spine and/or Temporomandibular Joint	44
Total	161

The spine was involved in 39 patients, the cervical spine being affected in 26 and the thoracic or lumbar spine in 13. In eight

patients, the temporomandibular joint was affected, including three whose cervical spine were also involved.

Symptoms and Clinical Findings: The symptoms and clinical findings, usually associated with juvenile rheumatoid arthritis, which were recorded at some time during the patients' illness, are listed in Table 7 in the order of frequency with which they appeared in the records.

TABLE 7—SYMPTOMS AND CLINICAL FINDINGS

		No. of Cases
Joint Inflammation		155*
Swollen	151	
Hot	97	
Red	65	
Fever		104
Joint and Bone Changes		100
Contractures		70
Spindling		68
Anemia		66
Anorexia		52
Adenopathy (lymph nodes)		44
Fatigue		40
Muscle Atrophy		39
Growth Disturbances		32
Cardiac Murmurs		29
Rash		29
Hepato-and/or Splenomegaly		24
Nodules, subcutaneous		23
Cardiac Damage		9
Epistaxis		7
Iridocyclitis		6
Colitis		5
Amyloidosis		4
Spondylitis		4
Purpura		3
Possible Lupus Erythematosus		2

*In 6 cases the records were incomplete.

Systemic Manifestations: For inclusion in the study, all patients had to have objective joint changes as described under criteria. In addition, other systemic manifestations were more commonly noted at some time during the illness of these children than would have been expected in an adult population. Sixty-five percent had a febrile response; anemia occurred in 41% and anorexia in almost one-third. Forty-four children (27%) had significant lymph adenopathy; and 24 (15%) had hepatic or splenic enlargement or both.

Other Manifestations: Several other manifestations appear worthy of comment.

Growth. Defects in normal growth and development were ob-

served in 32 (20%) of the patients. This group includes not only general gross underdevelopment and dwarfism, but also local growth defects involving the legs, feet, toes and jaw. This type of involvement was recently well described by Ansell and Bywaters.[1]

Eye Involvement. Iritis, uveitis and iridocyclitis have long been identified with rheumatoid arthritis. The severity of this manifestation in this series of cases was arresting. Of the six children who developed iridocyclitis, three had become blind in both eyes and one had lost sight in one eye. Edstrom, in 1947, noted three among 65 children with rheumatoid arthritis who had considerably decreased vision in one or both eyes.[1,2]

Cardiac Disease. Short, Bauer and Reynolds accept the fact that juvenile rheumatoid patients are more apt to have cardiac involvement than adults with this disease.[3] Pericarditis is considered to be the most common pathological manifestation. In the present series, over and above the 32 patients in whom cardiac murmurs alone were described (most of which were probably hemic in origin) there were nine patients in whom organic involvement of the heart was described. These effects ranged from pericarditis and cardiac enlargement to myocarditis.

Spondylitis. In connection with the four cases of spondylitis, it may be noted that recent investigators have emphasized that fusion of cervical certebrae may often be detected by x-ray when not suspected clinically. Coss and Boots pointed this out in 1946 and indicated that it was due, not to lack of segmentation (then the prevailing opinion) but rather to a fusion of the vertebrae secondary to the inflammation caused by the disease.[4] It is likely that such cases were not frequently sought for in the patients whose records are surveyed here.

Rash has been reported with relative frequency in juvenile rheumatoid arthritis although it is rare as a manifestation of the disease in adults. It is worth comment, therefore, that erythematous rash, not clearly defined, was present in 29 (18%) of the patients in our series.

Intensity of the Disease at its Peak: In 70% of the records the severity of the disease at its peak was described. Among these 112 cases, one half were classified as severely or very severely ill at some time in the course of their illness.

Fatal Cases: Six children died. In one of these cases the data were scant as the only case record seen was that of the child's final hospitalization which ended in death.

Among the five documented fatal cases were found two of the four cases of amyloidosis in the series, three of the nine cases of cardiac damage, one of the six childen with iridocyclitis (a child who had been blind for ten years) and three of the 24 cases of hepatosplenomegaly. These findings, therefore, would suggest a more serious prognosis.

TABLE 8—SEVERITY OF DISEASE

Recorded		112
Severe or Very Severe	56	
Moderate	32	
Mild	24	
Not Recorded		49
Total		161

Indicative of the unpredictability of the disease was the fact that one child, who was independent in everyday activities at the time of our home visit, despite some limitation of motion in the small joints and disturbed growth, died a few days later. Of possible relevance is the fact that she was on prednisone at the time of death. The four other fatal cases had moderate to severe deformity and required wheelchairs, braces or the help of another person in performing everyday activities.

THE CHILDREN'S CURRENT STATUS

To complete the data on the patient's current status, the investigator interviewed 79 patients and their parents in their homes. This section, therefore, is confined to 79 cases. Its data are based on the case records and on interviews with physicians, supplemented, in its evaluation of the children's performance of everyday activities (self-care, ambulation and ability to use public transportation) by the parents' descriptions of their children's activities and the investigator's observations.

Current Status of the Disease: Evaluations of the current status of the disease and of its peak intensity were obtained for 66 children. Of these, 41 (63%) were currently in remission. Of the children who had had arthritis for six years or more, three quarters were in remission at the time of study.

Current Performance of Everyday Activities: The children's functioning was recorded in terms of self-care, ambulation and ability to use public transportation. Currently, over half the children functioned normally. Of the 34 whose activities were limited, 20

required the help of another person in one or more activities, while 14, though independent, were limited by disease activity, fatigue, lowered stamina, awkward gait or poor physical development and, as a result, the tempo, extent and range of their activities were reduced.

TABLE 9—CURRENT STATUS OF DISEASE
as related to
Peak Intensity and Period Since Onset

	Current Status		
	Active	Quiescent	Total
	25	41	66
Peak Intensity of Disease:			
Severe	11	18	29
Moderate	11	11	22
Mild	3	12	15
Period Since Onset:			
1–5 years	15	9	24
6 years plus	10	32	42

Functional limitation correlated closely with current disease activity. Though all the children with active disease were limited to some extent in their daily activities, only six of the 46 patients currently in remission were limited; these six had all been severely ill in the past.

TABLE 10—CURRENT PERFORMANCE OF
EVERYDAY ACTIVITIES

	Performance Limited			
	Needs Another's Help	Does not Need Another's Help	Performance Normal	Total Patients
	20	14	45	79
Current Disease Status:				
Active	12	14	0	26
Quiescent	6	0	40	46
Not Recorded	2	0	5	7
Peak Disease Intensity:				
Severe	14	7	11	32
Moderate	5	2	15	22
Mild	0	3	12	15
Not Recorded	1	2	7	10
Period Since Onset:				
0–5 years	12	4	16	32
6 years or more	8	10	29	47

Physical Changes and their Social Implications: Even a child who performs all his everyday activities may have a physical change such as a stiff neck or an ankylosed wrist. Hence, the physical changes, i.e., the extent to which the disease has left its overt mark, were recorded.

TABLE 11—PHYSICAL CHANGES

Severe	8
Moderate	24
Minimal	31
None	16
Total	79

In Table 11, the classification of *severe* change includes children with marked and disabling physical effects, such as fusion of hip or spine, blindness or confinement to wheelchair or bed. Classified as *moderate* were such changes as a pelvic tilt, an underdeveloped jaw or contractures, which limit function and are socially recognizable. Finally, in *minimal* change, were included the children with slight limitations of motion of one or more joints who were not functionally or socially handicapped in consequence.

Thus, 32 children (two-fifths of those visited) had moderate or severe physical changes. Seven used wheelchairs, crutches or braces; eleven limped, lurched or walked stiff-legged; one was blind. Others had such visible effects as spindling of the fingers, marked stiffness of neck or wrists, a deformed jaw or generally underdeveloped growth.

Caution: The operative word through this section is *current*, i.e. the data picture the children's status when visited. It should be noted that after Tables 9 and 10 were compiled, a child there listed as in remission and functioning normally had a severe, incapacitating flare-up, a wheelchair-bound child began to crutch-walk and to anticipate walking unaided, and two children died.

ECONOMIC, EDUCATIONAL AND SOCIAL IMPACT

Juvenile rheumatoid arthritis was accompanied by serious economic, educational, vocational and social consequences for many of the patients.[5] As these factors frequently impinged on the child's medical care, they are briefly noted here.

Impact on the Child: Major interruptions of schooling were recorded for most of the children, caused by frequent, often protracted absences while sick at home or while hospitalized. All but six children were hospitalized at least once; half were hospitalized from three to 14 times. Half the children had had to depend on home teaching or special health classes for substantial periods of time, one quarter currently requiring such modified schooling. Moreover, one quarter of the school-age children were currently below their appropriate grade level as a consequence of their

arthritis. Of the teen-age children who needed special vocational planning because of the disabilities caused by arthritis, two-thirds lacked such help.

Serious personality changes—depression, anxiety, withdrawal, dependence, etc.—had developed at some time during the illness of 68% of the children visited; some of these currently persisted in 44%, though in many cases their severity seemed to be decreasing with physical improvement. About one quarter of the children appeared markedly dependent and passive in their home recreational activities.

Impact on the Family: Most parents faced serious problems in caring for their arthritic child. Heavy financial burdens were encountered by the great majority (estimated at $800 to $1000 annually) excluding the families who normally obtained all their medical care from municipal hospitals. This resulted in destruction of savings and debt (for over one third of the families). A number changed their residence, about a third turned to social agencies for financial aid and a few resorted to public assistance. The cost, in both money and time, of transporting the child to the source of medical care was also serious and long-drawn out for most of the families. Physically, the drain on the parents—especially the mother—of the sick child's care was severe and the emotional impact was also important, in four cases to the extent of requiring psychotherapy.

Almost without exception the families experienced serious, long drawn-out anxiety, and expressed the need for assistance in understanding the disease, its course and possible implications. In over half the group, the child's symptoms were so severe as to require continuous medical care while defying diagnosis for as long as a year, or even more. In this connection, it may be noted that half the parents used three or more sources of medical care for the sick child, one fifth of the group consulting from five to as many as 14 specialists.

The third major problem resulting from the disease was that of so planning the child's education as to ensure maximum continuity and content despite the frequent long interruptions and including, for the disabled teen-age child, timely and suitable vocational guidance.

Many parents were ill-equipped to deal with all these problems. In the cases where competent hospital social service was readily available, it proved helpful not only in obtaining direct and finan-

cial aid, but also in counselling, interpretation of the disease and in identifying the need for and obtaining vocational guidance. Only a minority of cases, however, seem to have been followed throughout their illness by social service.

Most of the parents, it was found, kept their child in treatment with the physician or hospital where the disease was diagnosed, even if this required added expense and considerable, difficult transportation problems. Moreover, whenever medical supervision was maintained beyond the point of apparent remission, this evidently provided valued support and reassurance to both patient and family.

Thus continuity of medical supervision and counsel, encompassing the full range of the child's medical, educational and developmental needs, appeared to be unusually important for this group of arthritic children.

Juvenile rheumatoid arthritis presents two special problems for the physician. Since it occurs infrequently, only a few physicians benefit from long-term experience with a substantial number of patients. The unpredictable, fluctuating character of the disease, the resulting anxiety to the parents, and the possibility that even serious handicaps may not be permanent, all complicate the physician's guidance of the parents, and his interpretation of the disease to them and to the child's teacher and social worker.

DISCUSSION

The current status of the 79 children visited by our investigator presents a relatively more hopeful picture than might be anticipated in view of the high proportion of children in the total group of 161 whose illness was severe at its peak.

Nearly two thirds of the children were in remission when visited and 40 of these 46 inactive cases performed everyday activities without restriction. Over half the 79 children functioned normally. Only 20 (25%) had functional limitations so extensive as to require the help of another person in performing everyday activities, the disease being currently active in 12 of these 20.

The majority of the children had had arthritis for at least six years. Significantly higher proportions of these children were in remission and functionally normal than among the children whose illness was of shorter duration.

These observations suggest that, at any one time, the majority of a group of arthritic children can function normally and that,

once the disease is quiescent, only a small proportion will be limited in function.[6] They further suggest that the longer the period since onset, the larger will be the proportion of children in remission and functioning normally.

Any such general picture, of course, would be modified by a number of factors in any individual case—the presence of irreversible joint damage or of such complications as cardiac damage, amyloidosis, iridocyclitis and, though perhaps to a lesser extent, hepatosplenomegaly. As reported by Barkin,[7] among others, the risk of sudden and serious excerbation is also always present. In our series this was illustrated in the case of the patient who, when visited by our investigator, was functioning normally, but who died a very few days later.

The nature of juvenile rheumatoid arthritis, moreover, is such that effects which are not strictly medical have great bearing on effective treatment. Almost without exception, the parents of the children in our series experienced acute and long drawnout anxiety and expressed the need for fuller discussion and explanation of the disease's implications than had been available to them. As this anxiety frequently affected their ability to care for the sick child adequately, the physician's ability to reduce it insofar as possible is important.

Arthritis also had serious consequences for the children's education; one quarter of the school-age children in our series being below their appropriate grade level as a direct consequence of their disease. The unpredictable course of the disease, and the consequent changes in the amount and type of teaching and recreation appropriate to the child, are unfamiliar to school officials or recreation agencies and may require special interpretation and more frequent action on the physician's part to ensure that the child's education and social development are as adequate as possible. The total absence of special vocational guidance and planning for the majority of the disabled teen-age children in our series was as regrettable as it was avoidable.

SUMMARY

Through a search conducted in cooperation with 85 hospitals in New York City, the records of 161 cases of juvenile rheumatoid arthritis, currently aged under 19, were obtained. Drawn from these records, data are presented on the patients' medical histories with some discussion of their recorded clinical symptoms. Six patients died.

The current status of 79 of the children was evaluated on the basis of home visits and conferences with the patients' physicians. Over half were functioning normally. Forty-six were in remission, of whom only six were limited in function. Over two-fifths exhibited physical changes characterized as moderate to severe.

The non-medical consequences of the disease—for both patient and family—were serious, involving the child's education, personality development and vocational potential and, for the parents, severe, extended anxiety and major economic burdens.

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CLELAND, J. B.: THE PYLORIC CANAL, URINARY BLADDER AND PERINEUM OF NEW-BORN INFANTS. (Medical Journal Australia, 45:420 Sept. 27, 1958).

The pyloric canal in the new-born infant is a very definite anatomical structure, clearly defined both externally and internally from the fundus above and very sharply demarcated from the duodenum below. On incising the pylorus, the thick muscle appears as a cartilagenous-looking layer attenuating upwards. The author suggests that the pyloric canal is so muscular at birth to enable the mucus and cell-debris that collect in the stomach before the baby is born to be passed into the intestine against the considerable pressure of the amniotic fluid.

The neonatal urinary bladder is surprisingly muscular and the author suggests this is so to enable the bladder to be emptied against the high infra-uterine pressure. The perineum of infants when the legs are slightly separated is usually nearly a plane surface, though a sulcus of varying depth may be present.

Other interesting features of the newborn are that the subcutaneous fat is lobulated, the cervical os is transversely rugose and ragged-looking, the cervical portion of the uterus is longer than the body, the epiglottis is usually folded, and the medulla and pons are firm and white (medullated) while the rest of the brain is almost gelatinous. M. A. B.

DEAFNESS PREVENTION IN CHILDREN

Loss of Hearing for the Middle Tones

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In a previous issue of the Archives (Vol. 75, No. 9) we stressed the loss of the low tones, the effect upon the child's ability to acquire and develop speech, and how it could be most effectively detected and beneficially managed. In another article (Vol. 75, No. 11) we stressed the effect of loss of the high tones from the standpoint of early detection, especially by the mother, and how the child can be helped. In this alert we will consider the loss of hearing for the middle tones, particularly those between 500 and 3000 cycles inclusive, the tones below and above this frequency band being heard normally or fairly well as determined by audiometric measurement.

This type of hearing loss is usually congenital. The patho-histological findings show a large destruction of hair cells on the basilar membrane in the middle turn of the cochlea, with little or no loss of these cells at the base or at the apex of the cochlea. In other words, there is no destruction of cells in the lower part or in the upper part of the basilar membrane. This is always a neural deafness and the audiometric curve is shaped like a "U", the upper parts of the "U" being near the zero line, or average normal, and the middle of the "U" showing losses varying from 40 to 90 decibels. If the loss is only 40 db. in these middle frequencies, the child hears the ordinarily loud voice—45 to 50 db in intensity—pretty well. But if the loss is much more than 40 db the voice has to be raised if it is to be heard when the speaker is more than 4 or 5 feet away from the child.

Children with "U" shaped congenital hearing impairments, though often labeled "slow talkers", do learn to speak quite normally and to understand language and speech quite well. The reason for this is two-fold: first, the child's low and high tone hearing abilities keep him alert to sounds in his environment; second, the child, hearing fundamental tones and perhaps some overtones, is often able to fill in the missing sounds mentally. Picture a jig-saw puzzle in which the right and left sides are in place but in which

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the bottom center is sketchily filled in. Enough of the picture may be showing to give you an idea of what it will look like when complete. Likewise with hearing—with good recognition of the frequencies above 2000 and those below 500 the child will be able to fill the frequencies in which he is deficient, often with remarkable accuracy.

Early detection of "U" shaped hearing loss curves is particularly difficult in young children. Neither pediatrician nor parent may have reason to suspect hearing loss. The pediatrician usually has no history of ear disease and no obvious pathology to guide him. Both parent and pediatrician have evidence that the child hears even some faint sounds like a cat's purr (low) or a bird's chirp (high) from a normal distance, and is therefore alerted to pay attention to whoever is speaking to him and begin to augment his hearing difficulties by becoming a good lip reader. The child's speech is understandable and his vocal language and understanding can be remarkably good.

Add to these barriers to detection another obvious difficulty—that of obtaining precise measurement of any very young child's hearing. Even if some nagging doubt were to exist in either parent or pediatrician's minds it might be difficult to get a busy otologist interested in the case because there is probably no treatment that he can offer for this type of hearing impairment plus the fact that otological scientists have had little opportunity to study these cases of middle tone deafness due to the reasons we mentioned above regarding early detection. There is very little literature on the subject.

So it is not unlikely that the "U" shaped hearing problem may remain undetected until the child enters school. And many schools do not test kindergartners unless they "seem to have trouble in hearing or speaking".

By the time the child has reached the first grade he has often developed considerable lipreading skill to in part compensate for his hearing loss and he may therefore be overlooked once more by the school authorities unless he has difficulty in learning to read. The hearing loss probably will not be really recognized until the child is seven or eight years old.

At this point the pediatrician enters the picture in a most important role. He will first of all accept the school's case finding and carefully investigate. He will than have to inform the parents and help them to accept and understand their child's need for spe-

cial help—his need for lipreading, speech and auditory training and in many cases a hearing aid. Hearing aids may be of limited benefit for this type of hearing loss but should be considered and tried.

We believe there is a need for both the otologist and the pediatrician to familiarize themselves with the public school programs for hearing testing and to support them actively with a view to improving the case finding surveys in their community.

In closing this series of articles on the three specific types or classifications of deafness—low tone, high tone and middle tone losses—we would like to again suggest that the pediatrician seek information about community resources for his young hearing impaired patients from the local medical society, hearing and speech clinics in hospitals and universities, and from the American Hearing Society and its Member Agencies like the New York League for the Hard of Hearing in New York City. They can give specific help regarding hearing aid evaluation, hearing and speech therapy and parent counseling.

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TAYLOR, G. D.: *Complications of Middle Ear and Mastoid Disease Despite the Antibiotics.* (Southern Medical Journal 51: 1089 Sept., 1958).

A series of 86 cases of suppurative mastoid disease managed surgically during an 8 year period is reported. The onset of the disease occurred in 50% of the cases after the introduction of the antibiotics. In all cases the patients were treated with sulfonamides or antibiotics at some time before operation. The disease process progressed despite these drugs, and surgical intervention was necessary. Few modified radical mastoidectomies were done because of the far advanced process in the mastoid and middle ear. Forty-one complications occurred in 27 cases, or 31% of the series. Early surgical intervention would have prevented most of these complications. Cholesteatoma occurred in 65% of the cases.

Thirteen cases are described. Among them are 9 cases of far advanced cholesteatoma which illustrate the futility of delaying mastoid operation when it is indicated. In one case of secretory otitis media and mastoiditis, there was response only to surgical treatment of the mastoid. Three cases of acute suppurative mastoiditis which developed despite the antibiotics are presented. One of these was complicated by facial paralysis. AUTHOR'S SUMMARY

SCROFULA: THEN AND NOW

1500 Years of Tuberculous Lymphadenitis in Accessible Nodes

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Scrofula, derived from the Latin, is the diminutive of *scrofa* meaning breeding sow, an animal characterized by a large neck comparable to the swollen neck of one who suffers from tuberculous cervical lymphadenitis. As such, the term should be limited to cervical lymphadenitis but Wiseman[1] (1622-1676) also includes other lymph nodes in his classical description of the disease and, certainly, tuberculousis of the axillary and inguinal nodes is in the same category as the more important cervical lymph node tuberculosis. Scrofula was also known as the king's evil and hence was believed to be amenable to the king's touch.

This form of treatment, according to Gale[2] was practised by Clovis, the first Christian king of the Franks (481-511). In England, Edward the Confessor (1042-1066) used the king's touch with great success (Wiseman[1]) and Charles II cured many thousands. (Wiseman, the court physician, could be prejudiced!) Nevertheless, the treatment was discontinued by Queen Anne (1702-1714) but not before she had tried it on Samuel Johnson. Her failure with that literary giant, then a young child, was probably unknown to her and could not have been her reason for abandoning a form of treatment which had been popular for 1200 years.

Any therapeutic measure employed for that length of time must have been followed by improvement or cure in some cases. Otherwise, it would have been abandoned even though it was associated with ceremonies of a religious nature. This, in itself, is evidence that tuberculous lymphadenitis has a tendency to spontaneous improvement or resolution, a fact which should not be forgotten when considering the efficacy of any treatment.

In spite of this, scrofula was a common disease and presented a serious problem. Its etiology was little understood by the medical profession but, in 1885, Allbutt[3] noted its similarity to tuberculosis and suggested rational treatment. Of course, some form of medication had to be used such as cod liver oil, mercury and the syrup of ferous iodide but he did stress the value of hygienic surroundings, of adequate diet and of fresh air and sunshine. He also advocated

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the radical surgery practised by Teale[3]. This consisted not only of incision and drainage but also removal of the diseased nodes where possible and of curettage of caseous areas. By our present standards this would not be considered radical but it was much more extensive than any other contemporary surgery used for scrofula, and likewise more effective.

Around the turn of the present century Dowd[4] advocated much more extensive surgery which aimed to remove all the diseased nodes. His results were the best up to that time but the incidence of tuberculous lymphadenitis was decreasing and the pendulum swung back to means other than surgery.

This was particularly true in England where Thompson[5] vigorously opposed surgery and advocated only the medical treatment noted above plus tuberculin vaccination although this method required months or even years and in many instances was a failure. Barrington-Ward[6] and Evans[7] felt that surgery was necessary to effect a cure in any but the cases of minor extent after the patients had a long preliminary treatment with fresh air, nourishing diet and rest. Rest was carried to the extent of encasing the neck in a splint or plaster collar. When these gentlemen did get to surgery they employed the painstaking procedure described by Dowd and were careful to preserve the eleventh cranial nerve which supplies the sternomastoid and trapezius muscles. These twentieth century surgeons recognized the probability of tuberculosis elsewhere, especially in the lungs, but they did not stress its relation to the lymph node disease nor the importance of treating the pulmonary disease with the lymphadenitis. Of course, the long preliminary hygienic treatment *was* treatment of the pulmonary infection whether or not they recognized it, and it should not be relegated to the status of the "king's touch."

On the other hand, ultraviolet radiation, which had considerable popularity at the time, could be classed with the "king's touch." The treatment took weeks or months and any good results could be ascribed to the natural course of the disease just as well as to the treatment.

Roentgen ray treatment is in a different category. This was widely used at that time and still has its advocates. The swellings subsided fairly promptly and sinuses healed, at least temporarily. However, in a few instances wide spread dissemination of the infection did occur during the treatment and recurrence was frequent. Because of this and also because the hazards of radiation have recently come into prominence, x-ray therapy can well be discon-

tinued in the treatment of scrofula, except in rare instances. Other forms of treatment are less dangerous and more effective.

Meanwhile much was being learned about the pathology, bacteriology and mode of transmission of tuberculosis. The work of Medlar[8] was of paramount importance in the pathology of the disease and also in understanding the effect of chemotherapy on the tuberculous process.

Tuberculosis of the superficial lymph nodes has a varied background. It may be part of a generalized miliary tuberculosis. In this instance the patient is febrile and has the other manifestations of miliary tuberculosis.

It may be associated with an active pulmonary lesion, which is usually more than a primary complex, and has fever cough and sputum. Or it may be concomitant with a healed or quiescent lesion, often of long standing.

It may be associated with extra-pulmonary disease such as Potts disease or other bone, joint or visceral infection. This is rather infrequent.

It may be present without any demonstrable lesion elsewhere, and this is rather common. Samuel Johnson probably had such an infection. His biographers tell of his years as a hack writer when he was ill-fed, ill-clothed and ill-housed, conditions which would have caused a pulmonary lesion to flare up into fatal activity and deprived English letters of one of its most colorful characters.

Tuberculous lymphadenitis, in the absence of lesions elsewhere may have several portals of entry. Medlar[9] believed that the infection in the upper cervical nodes entered through the tonsils or adenoids. Riley[10] has demonstrated that the infection can enter the lymph nodes by way of the lungs without causing any pulmonary infection. Lymph nodes also become infected from drainage of tuberculous skin lesions and I have seen one instance of tuberculous nodes associated with tuberculosis of a tear duct.

The organism of bovine tuberculosis was once prominent in lymph node tuberculosis and still is in some countries. In 1908 Dowd[1] reported that Park found about 30% of his cases to be of bovine origin but at that time pasturization of milk was rare and tuberculin tested dairy herds almost unknown. Now, in the United States the strict supervision of the milk supply has about eliminated bovine tuberculosis and hence, the human organism is responsible for almost all tuberculous lymphadenitis.

The human disease is also much better controlled. Therefore, the incidence of scrofula is much less although it is still present and

certainly not rare. The impact of antituberculosis chemotherapy has further served to lessen but not to eradicate it. Before discussing the chemotherapy consideration of the pathology of tuberculosis is in order because it is this which dictates the use of the drugs.

Tuberculosis starts as an acute infection and the picture is that of an acute inflammatory process with edema, polymorphoneuclear leucocyte infiltration and necrosis. Then the necrosis becomes caseous, round cells replace the leucocytes, giant cells appear and tubercles are formed with surrounding fibrosis. The process may stop at this point with later calcification of the caseation. More often, however, the caseation necrosis liquefies and a cold abscess forms. If this is in the lung the cold abscess breaks into a bronchus and the pus is coughed up. In a lymph node spontaneous drainage occurs only after the cold abscess has worked its way through the surrounding fibrosis and perforated the skin. The resulting sinus may drain for months or years because the caseous node is at the bottom of it and acts as a foreign body. Occasionally the entire caseous area liquefies and when this has drained out the wound heals. Unfortunately this felicitous circumstance is rare. Eventually when the wound does heal there is an irregular, unsightly scar.

The clinical signs vary with the pathological condition. In the early, acute phase the swollen lymph node is tender and there is frequently a temperature elevation. If this is associated with miliary tuberculosis the patient is acutely ill and presents all the other signs that go with miliary tuberculosis. If the infected lymph node is associated with the active phase of pulmonary tuberculosis, the lung infection dominates the picture. If the pulmonary lesion is healed, quiescent or absent the symptoms and signs are limited to the involved nodes.

After a time which varies from a few days to a month or more the fever and tenderness are no longer present but the swelling persists. If caseation has not occurred this swelling will gradually disappear. In the presence of caseation the swelling persists but may not increase or may even diminish as the non-caseous areas fibrose or absorb. If, as usually happens, liquification takes place tenderness recurs, swelling increases and becomes fluctuant, the skin reddens and perforation occurs. This late tenderness which is caused by a cold abscess must not be confused with the initial tenderness which is the result of acute inflammation.

Because scrofula can be confused with Hodgkins disease, lymphosarcoma or other lesions affecting lymph nodes, biopsy may

be necessary for diagnosis. When this is done the entire group of nodes exposed should be removed because in the presence of tuberculosis a node only partially removed is apt to produce a cold abscess which will reopen the wound and develop a chronic sinus.

The effect of therapy varies with the pathological state of the disease. X-ray therapy shrinks lymphoid tissue and produces fibrosis. It does not sterilize the infection and it has no effect on caseation necrosis. If the fibrosis has not permanently limited the disease the swelling recurs and the lymphoid tissue reappears.

On the other hand, chemotherapy in the form of streptomycin, isoniazid and PAS *does* attack the infection. In the acute phase it is highly effective and may terminate the disease at this point. In this it differs from radiation therapy which may cause diffuse dissemination when used in the acute phase. When fibrosis occurs, and especially in the presence of caseation, the penetration of the drug is limited and usually ineffective. However, since isoniazid, in some instances, seems to hasten the formation of a cold abscess this drug probably does penetrate more than the others. With the formation of a cold abscess the drugs are effective in reducing the infection in the newly contaminated tissues. The drugs are used in treating lymphatic infection in the same manner and dosage as they are in the inaccessible pulmonary infection and the organisms develop resistance just as rapidly.

From the foregoing it is evident that the therapy of tuberculous lymphadenitis depends upon many factors and cannot be stereotyped. In the presence of active pulmonary disease treatment should be directed toward the pulmonary condition and this treatment is also that of the scrofula. Rest, in a sanatorium, if possible, with adequate diet should be associated with chemotherapy. As the pulmonary disease is controlled the lymphadenitis may disappear. If it persists after the pulmonary disease is controlled but not cured surgery should be considered for the nodes. Removal of the diseased nodes has a favorable effect on the pulmonary disease.

When the active disease is limited to the lymph nodes a different situation exists. In the early acute phase, especially in the presence of fever, rest is as necessary as in any other acute infection. At this time chemotherapy is most effective and should be employed for a month or six weeks, or longer if there is marked decrease in the swelling. If the scrofula comes under treatment after it has been present for some time and tenderness no longer exists chemotherapy is indicated and activity should be somewhat curtailed, but, in the absence of an active pulmonary lesion sanatorium care is an

unnecessary expense. If the swellings show little or no change after six weeks of chemotherapy caseation is certainly present and surgery is the procedure of choice. Chemotherapy continued under these circumstances is in the class of the "king's touch." Should a cold abscess form earlier, surgery should be undertaken promptly because a draining sinus complicates the treatment.

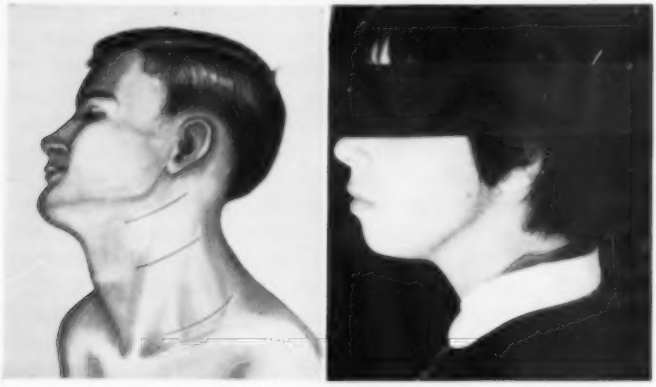

Figure 1. Incisions in neck for tuberculous nodes. These give adequate exposure and heal with inconspicuous scar.

Figure 2. Scars in neck of 14 year old girl operated upon for tuberculous nodes. Three operations were done through the upper scar. The first was for biopsy; it drained many months and left an unsightly scar. In each of the next two operations the preceding scar was excised. In the last operation a cold abcess was present and each of the last two operations healed by first intent. Prior to the second operation the patient had repeated x-ray treatments and took isoniazid and PAS for two years. The node removed in the last operation appeared and broke down while the patient was under chemotherapy. The lower scar was from a second incision made in the second operation.

Surgery for tuberculous cervical lymph nodes implies the removal of all the diseased nodes with the preservation of all the important structures, especially the eleventh cranial nerve. The incisions are made in or parallel to natural creases in the neck so that when they heal the scars will not be noticeable. (Fig. 1) The technique has been described elsewhere[11,12] and will not be repeated here. It is not the radical neck dissection used for cancer which sacrifices important structures for the sake of thoroughness. The appearance of another tuberculous lymph node, something which occasionally happens, does not imply failure and the node can be removed through the same scar. The wound heals with an inconspicuous scar quite unlike the unsightly scars resulting from the spontaneous drainage of a cold abscess. (Fig. 2)

Contamination of the wound by tuberculous pus can be disregarded. Should there be a large cold abscess however, drainage by a small incision and subsequent treatment with proteolytic enzymes is advisable. This may be adequate but, if not, subsequent operation is easier and more effective after such drainage.

These same principles are equally applicable to nodes in the axilla and groin except that the incision can be made in the most advantageous place since usually it will not be visible.

Chemotherapy may be continued for ten days or two weeks after operation to cover the surgery, although, in my experience, it has not prevented activity in a node which has been missed at operation. Of course, tuberculous lesions other than those in the lymph nodes should have the same chemotherapy after operation as before.

SUMMARY

1. Scrofula, i.e., tuberculous lymphadenitis in neck, axilla and groin, has been in the public consciousness for at least 1500 years.

2. It has been subjected to many forms of treatment, among which the "king's touch" was in vogue most of the time.

3. In judging the worth of treatment it must be remembered that spontaneous improvement and cure can take place in due course.

4. Treatment depends upon the presence or absence of other tuberculous lesions and in all cases should be initiated by general hygienic measures and chemotherapy.

5. Surgery is an effective and safe form of treatment. It should be employed when chemotherapy has not produced an adequate result and not unduly delayed in the absence of tuberculosis in other organs.

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NORETHANDROLONE* AND RESERPINE

A Study of Treatment of Nervous, Underweight Children

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A recent study by Brown, Libo and Nussbaum[1] indicated that the anabolic agent norethandrolone*, a synthetic steroid, safely and effectively restored appetite and increased rate of weight gain in chronically underweight children. Many of the children in this study who responded favorably to norethandrolone had failed to show any clinical improvement with vitamin B_{12} and other agents having an antianorectic effect. It is probable that norethandrolone initiated better weight gain than did vitamin B_{12} in these children since it is reported to exert a potent protein anabolic[2] as well as an appetite-stimulating effect.

Other recent studies[3,4] have pointed out the value of reserpine combined with the principal antianorectic vitamins for the treatment of underweight children in whom this condition was associated with nervous hyperexcitability. It was shown in these studies that reserpine safely and effectively controlled underlying emotional disturbances in these children which at least in part accounted for their nervousness as well as their stubborn refusal to eat properly.

The consideration that norethandrolone with reserpine would constitute a highly effective combination in the medical treatment of such children prompted us to employ it in twenty-five boys and twenty-two girls, ranging in ages from 1 year 11 months to 13 years 6 months. At the same time we administered norethandrolone alone to fourteen boys and nine girls, ranging in ages from 6 weeks to 17 years.

All of the children had failed to gain weight normally due principally to poor appetite, and those receiving reserpine in addition to norethandrolone were of the nervous, "fidgety" type. Additional findings in twelve of the children were acne, chronic eczema, enlarged tonsils, celiac disease, fecal impaction, malaise, catabolic state resulting from corticosteroid therapy, adrenopathy in addition to recurrent parotitis, and cryptorchism associated with hypopituitary cachexia and dwarfism.

Dosage of norethandrolone varied with the age and response of the patient and ranged from 5 mg. every other day to 10 mg. three times daily. Reserpine dosage for all patients receiving the drug

*Nilevar®.

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was 0.05 mg. three times daily. Therapy was continued for from one to eighteen weeks, and was interrupted for one month in two children and for two and three months in two other children. Six children were examined at periods of from one to three months after the medication was discontinued.

In determining response, we considered overall improvement of the child more important than absolute increase in rate of weight gain alone. The chemical similiarity between norethandrolone and testosterone warranted our close observation of each child for the possible development of androgenic side effects, even though previous studies[1,5,6] have shown an exceedingly low incidence of these effects in children.

RESULTS OF THERAPY

All of the children showed a substantial weight increase while on norethandrolone. It was our impression that this increase was accompanied in most cases by improvement in appetite, disposition, vitality and general appearance as well. In addition, the children who also received reserpine became less nervous and irritable, at the same time showing better behavior and temperament.

It was our further impression that norethandrolone therapy in no instance induced weight gain beyond the child's normal constitutional limit. The rate of weight gain observed in six children one to three months after therapy had been discontinued was diminished slightly from that established during treatment, but still was notably better than it had been before norethandrolone was started.

Especially noteworthy was the good weight response and general improvement observed in the children with weight loss due to organic and pathologic disorders. For example, a 7 year, 6 month-old boy whose failure to gain weight properly was associated at least in part with rheumatic heart disease was given norethandrolone 20 mg. daily and reserpine 0.05 mg. three times daily for four weeks. Medication was discontinued for one month and then reinstituted for another month. During both courses of therapy the child showed a marked increase in rate of weight gain which continued to the time he was last seen, four months after therapy had been discontinued. At this time the child showed a gain of 10 pounds, 12 ounces since the start of therapy, and in addition he looked better and was less irritable. The additional weight gain caused no signs of cardiac embarrassment.

In a few instances some of the weight increase induced by noreth-

androlone was considered due to sodium and water retention. However, most of the children showed a weight increase attributable principally to increase in protein tissue.

Androgenic side effects occurred only in one child, a three-year-old boy who experienced occasional priapism while receiving norethandrolone 20 mg. every other day for two weeks. Side effects attributable to reserpine were observed in three children, two of whom experienced sleepiness and nasal stuffiness. No toxic effects from either drug were observed. Since norethandrolone might be considered contraindicated in patients with acne due to its slight androgenic activity, it was interesting to observe that acne in a 17-year-old boy improved notably during therapy with the drug.

DISCUSSION

Anorexia in children often results from underlying emotional disturbances incurred in infancy which stem from such factors as forced feeding, and later from overly strict "table training." Often the most effective treatment of such children is the re-education of the parents. When this is too late to do any good, both psychologic and medical treatment must be directed toward the child.

Many such children encountered in practice present a picture of nervous hyperexcitability, manifested by "fidgetiness", lack of cooperation and generally poor disposition. In addition, they almost always are underweight and poorly nourished due to their stubborn refusal to eat properly.

The medical treatment of these children is directed at attempts to relieve the underlying emotional disturbances while simultaneously encouraging a better dietary intake to correct the resulting underweight condition. Recent studies[3,4] indicate that the mild tranquilizer reserpine combined with the principal antianorectic vitamins effectively fulfilled these dual goals of therapy.

It is reasonable to expect that testosterone or one of its compounds would initiate weight gain in these children even more effectively than vitamins, since it exerts a potent protein anabolic as well as an appetite-stimulating effect. This additional anabolic activity would make it worthy of trial especially in refractory cases which failed to respond to vitamins. However, it is widely recognized that testosterone also exerts an androgenic activity capable of producing virilization as well as of inducing premature epiphyseal fusion, which makes its use in children dangerous and therefore largely impracticable.

SUMMARY

In the present study, seventy anorectic, underweight children ranging in age from early infancy to late adolescence were treated with the anabolic agent norethandrolone (Nilevar®), a synthetic steroid. Forty-seven of these children simultaneously received the mild tranquilizer reserpine to control nervous hyperexcitability. Of the seventy children, only one showed a slight, transitory androgenic response. This finding supports previous reports[1,5,6] that norethandrolone possesses an exceedingly small androgenic activity and therefore is safe for administration in children. The gratifying weight increase observed in the majority of the children in the present study supports another previous finding[7] that norethandrolone given by mouth could be expected to exert at least twice the protein anabolic activity of testosterone.

In addition to weight increase, the majority of the children showed a marked improvement in appetite, vitality, disposition, general appearance and study and sleeping habits as well. The forty-seven children who also were given reserpine to control nervous hyperexcitability generally showed less irritability and became more cooperative and better disposed. Water retention, which was noted in a small number of the children, was not troublesome. No toxic effects from either drug were observed.

On the basis of our observations in the present study we employ norethandrolone almost routinely to hasten the convalescence of hospitalized children. We have found that its administration in these children rapidly restores their weight and strength, thereby permitting us to send them home sooner. The present results also have prompted us to employ norethandrolone in premature infants to hasten weight gain, although to date we have made no conclusions concerning this use of the drug.

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Clinical Conference

ST. LUKE'S HOSPITAL, New York

September 23, 1958

J. FREDERICK EAGLE, M.D.
Director of Pediatrics, presiding

UNDESCENDED TESTICLE

DR. EDWARD J. ARIDA, Intern: P. M. (SLH #339-423) is a six year old boy who was admitted five days ago with a chief complaint that he had but one testicle. On admission he was a normal, healthy, six year old boy, his only defect being a right undescended testicle and a right inguinal hernia. He was operated on the following day. The testis was freed and attached to the fascia of the thigh, and a herniorraphy was performed.

DR. EAGLE: The problem of the undescended testicle is one that in many instances is quite controversial. Not infrequently, we find pediatricians taking one point of view, pediatric surgeons taking another, and urologists taking a third. We have all three disciplines here today so let's start with the pediatric surgeon, Dr. Edward Stanley-Brown.

DR. STANLEY-BROWN: There are several varieties of undescended testicle. The testicle is sometimes completely intra-abdominal. It can also be ectopic, down in the perineum. The common location for the undescended testicle is in the inguinal canal. The point about a true undescended testicle is that it cannot be moved from its position.

There is another variety which is referred to as a retracted testicle or a migratory testicle which is though to be due to an overactive cremasteric muscle. In examining such a child, press down along the spermatic cord. The testicle will come down into the scrotum; you can grasp it and pull it down where it belongs. These patients do not have a true undescended testicle and this condition will correct itself at puberty. I believe this is also the sort which will descend with the administration of gonadotropic hormones and do not need to be operated on.

In the management of the true undescended testicle, as Dr. Eagle has indicated, there is a great deal of controversy as to what age the child should be before surgery is undertaken. To give you an idea of how widespread the different thoughts are, there were two

articles in the *Journal of the American Medical Association* last year. The first one by Dr. Kimbrough, a urologist, recommends that surgery be done between the ages of three and six. Dr. Everett Koop in Philadelphia recommends that it be done before the age of six. Dr. Robert Gross, who with Dr. Theodore Jewett, reported on 1,222 children, feels that between 9 and 11 years is the proper time. Dr. Orvar Swensen in his excellent new book on pediatric surgery prefers the age of ten or eleven. The other article in the *AMA Journal* by Dr. Drake recommends waiting until the patients are sixteen or seventeen. I don't know what the right age is. Perhaps Dr. Taylor can enlighten me. I think that from a technical point of view it is perhaps easier to do it when the child is around eight or nine years old. About 90% of them have an associated hernia and certainly this should be repaired at the same time. There is always the problem of fertility and in this regard Dr. Gross has sixteen patients who had bilateral undescended testes. He reported that of these 79% grew up, got married, and had children.

The Torek operation has been mentioned in connection with this case and I am interested in knowing the reasons for using that procedure. It seems to me that that is making two operations than can be done in just one.

DR. EAGLE: Thank you Dr. Stanley-Brown.

DR. JOHN TAYLOR, Director of Urology, St. Luke's Hospital: At the Hospital for the Ruptured and Crippled there was a theorem which I think we proved pretty conclusively and that was practically all undescended testicles are ectopic. I have also seen them in the femoral canal, Dr. Stanley-Brown, in addition to the other areas you mentioned.

The other theorem postulated was that all undescended testes had hernias which required a special effort to demonstrate. Hernial sacs in these little children are just like wet tissue paper. Even when you have the sac open and see the vessels on the ouside it looks as if there is nothing between you and the vessels. We believe that these testicles are ectopic and that they are all associated with congenital hernias.

We saw many cases that had migratory testicles. The children are sent in by their family doctor for undescended testicle, and as Dr. Stanley-Brown said, you can milk it down into the scrotum. I think that one of the first things that should put you wise to the fact that you are not dealing with an undescended testis, is that the scrotum is developed. You never get a fully developed scrotum with a

true undescended testicle. If you see a boy with a normal scrotum, with a testicle in one side but not in the other you can be pretty sure that it is migratory and can be pushed down. We also believe that hormones do not bring down any undescended testicle. What they do bring down are these migratory testicles.

Before the Torek operation was described, we found that six months or a year after placing the testis in the scrotum, it would have retracted. I do not believe that the cord can be freed up enough so that the testicle can be placed in the scrotum and stay there without fixation. Some use rubber bands but we used the Torek procedure. The testicle is pulled down through the scrotum and attached to the thigh. It is left there for a year and the boy is allowed to play ball and do everything else. Abduction of the thigh stretches the cord, and a year later when the testicle is detached, you have a testicle that will stay down. I know of no other operation that takes the place of the Torek unless it be traction and that is only for a short time.

Robinson and Engle did a very interesting piece of research showing that up to six years of age there is no development of the testicle, and from six to ten there is some, but not much development. From age ten on it grows and, of course, at puberty it fully develops. Now if that is true, and I think they proved it rather conclusively, I would agree with Dr. Stanley-Brown that eight years is the best time for surgery. If the testicle is developing you want it down where it can develop. The scrotum is a thermostat, it expands in the heat and retracts in the cold to keep the temperature at a constant level. It was brought out years ago that there is only one degree difference in the heat between the scrotum and the inguinal canal. It was shown that one degree of heat would prevent the testicle from developing, that an undescended testicle in the groin could not develop. There is at least two or three degrees difference in the abdomen.

To recapitulate, I think that almost all patients with undescended testicles have hernias, practically all of them are ectopic, that hormones do no bring down a true undescended testicle, and that the ideal time to operate is between eight and ten years.

A final problem concerns the incidence of carcinoma in the undescended testicle. This past summer we removed an intra-abdominal testis which could not be placed even in the inguinal canal. In another case I advised leaving the testis alone. Everybody who writes a book just looks at a previously written book and repeats that 2% of all undescended testicles are carcinomatous. However,

authentic case reports are hard to find. Mostofi who is one of the finest pathologists in the country at the Armed Forces Institute in Washington feels that he can substantiate the increased incidence of malignancy in the undescended testis. So I think if I had the abdomen open and came across an undescended testicle I would take it out just on the chance that it might become carcinomatous.

DR. EAGLE: I admit I was quite concerned about this patient who had the orchidectomy this summer and I spoke to Dr. Taylor about it. I was very dubious about this malignancy business because the incidence of carcinoma of the testes is 13 per 100,000 and the incidence of true undescended testes that is either intra-abdominal or ectopic is of the order of one in 500. On the other hand, the same article reports 700 patients with proved carcinoma of the testes, 10% of which were undescended. If those figures are correct and comparable with the ones at the Armed Forces Institute I think that maybe I will have to revise my thinking and if the testis is intra-abdominal and you can't get it at least into the inguinal canal that the thing to do is to take it out.

DR. TAYLOR: It will never be any good anyway.

DR. EAGLE: Which brings up another question. Is there any point in bringing any of these ectopic testes down for the purpose of fertility? There is certainly disagreement on this. Many people feel as Dr. Taylor does that these testes are never going to be any good. On the other hand, Jewett and Gross report thirteen patients operated on for bilateral undescended testes who are presumptive fathers.

DR. TAYLOR: Thirteen out of how many?

DR. STANLEY-BROWN: 1,022. Seven actually were fathers, the other six had high sperm counts.

DR. EAGLE: True, but not until adolescence. If chorionic gonadotropic is given to a five year old with migratory testes, they will descend in a matter of weeks.

DR. TAYLOR: I don't think so, but that is only my opinion and I could be wrong.

DR. EAGLE: Engle's original work was in monkeys and he showed that in monkeys the testes are apparently not descended until they become pubescent. In preadolescent or prepubescent monkeys, gonadotropins would stimulate descent of the testes.

DR. TAYLOR: True, but here one is dealing with a normally placed testicle, while the cryptorchid in the human is ectopic—how much gonadotropic and over how long a period?

DR. EAGLE: I haven't read that original article which was written thirty-five or forty years ago. It was from those studies that gonadotropins were given a clinical trial. In an article in the *Journal of Urology* two or three years ago Engle felt that the prolonged use of chorionic gonadotropin was a mistake and his recommendations were to use four to five thousand units daily for three days and if the testes were not down within a week to operate. He objected to larger doses for a long period of time which then produced secondary changes; an enlargement of the phallus, and endocrine disturbances. But he had no real objection to rather large doses for a short period of time under those circumstances where there was a possibility that the testicle might descend. A second argument is that by using chorionic gonadotropin the size of the cord and other structures is enlarged making surgery a lot easier.

DR. TAYLOR: Well, there is nothing difficult about operating on an undescended testicle, even with the smallest cord if it has the necessary length, and that is the important thing. Of course, the great problem is to get them down into the scrotum.

DR. EAGLE: In any event, both Dr. Taylor and I agree that we would not use chorionic gonadotropic although for different reasons. At this point I might add that testosterone should never be used, since this will prevent any maturation of the testes by inhibiting the production of the patient's own gonadotropins by his pituitary.

DR. LEO F. J. WILKING, Pediatrician: I would like to know what percentage of undescended testes can be put into the scrotum?

DR. TAYLOR: I think you can get them all in. I remember a patient we had here in the ward years ago who had bilateral intra-abdominals, I could bring them down only to the spine of the pubis. A year later in a second operation I was able to perform a first stage Torek and in a third procedure I did a second stage Torek. I've never cut the epigastric vessels to bring them down. Some people claim that if you do this you get additional length but I think that if they are so short they won't go all the way down to the thigh it is better to fasten them to the spine of the pubis as a preliminary procedure.

DR. WILKING: Do you think that intra-abdominal testes should be looked for?

DR. TAYLOR: Yes, just in case they might be just inside the internal ring. If they are farther away you never will get them into the canal.

DR. WILKING: Is there any way you can tell that they are just inside the ring without operating on them?

DR. TAYLOR: I don't believe so. I've never been able to feel an abdominal testicle. They are pretty small.

DR. WILKING: So that at least you should look in the ring and see if you can find one?

DR. TAYLOR: Yes, but I believe that the incidence of successful results is rather small. I think most undescended testicles are atrophic and are not going to function. But you cure the hernia and give it a chance.

DR. CATHERINE LODYJENSKI, Pediatrician: If you have a patient where you don't clinically find the testicle, is it justifiable to try a short course of hormones before you do any surgery?

DR. TAYLOR: No, if you can't find a testicle, no hormone can affect a testicle that is intra-abdominal—turn it around, and bring it down through the canal.

DR. EAGLE: I would like to ask you one other question in regard to hormones. There certainly is considerable disagreement in the literature about their use, but I think you would agree that a migratory testicle will descend if chorionic gonadotropic is given.

DR. TAYLOR: I don't agree with that. If they are migratory they are going to come down anyway.

FRANKLIN, A. W.: Iatrogenic Disease in Childhood. An Essay in Definition. (British Medical Journal, 5095:559, Aug. 30, 1958).

So we see that all the powers mobilized for healing—the drug, the prophylactic, the apparatus of diagnosis, the hospital, the outpatient clinic, the ancillary personnel—all carry also the power to damage. And in the end of it all the doctor and the patient face each other, two human beings prone to error, both with mankind's portion of fantasy and fear, one trying to be scientific, the other to be brave, each trying to fathom the other by question and answer, but with the help of instinct and feeling as well as words. Small wonder that at times the patient is left the worse for the encounter with "a harmful disorder unwittingly induced in him by the inappropriate use of drugs or faulty management or treatment or because of the physician's examination, manner and discussion" or with, in two words, iatrogenic disease. AUTHOR'S SUMMARY

To the Editor
Archives of Pediatrics
New York, N. Y.

April 7, 1959

In a recent issue of the ACHIVES OF PEDIATRICS, *the writer's paper on listeria meningitis in infancy was published.*[1] *Considerable reader interest was demonstrated in this paper, as evidenced by reprint requests, correspondence, etc. Several requests came from behind the Iron Curtain, particularly East Germany, where epidemics of human listeriosis have been reported. Interesting letters have also been received from communicable disease centers, and veterinarians.*

Because of the marked interest shown in this subject, it may be worthwhile to report some additional information not available at the time my original paper was published.

Readers may recall that I reported two cases of listeria meningitis from the Oakland, California area. Both infants were negro neonates, and the outcomes were fatal. In the same year, two additional cases were reported from the Oakland area, and both of these infants were negro neonates. One infant survived, and the case was published.[2] *The other case was not published, but was learned of through personal communication.*[3] *In this latter case, despite prompt recognition of the organism, and comprehensive therapy, the infant died.*

Because of the obscure epidemiology and uncertain cause of infection in human listeriosis, much remains to be learned. Animal-to-man transmission has been predicated, but strong evidence is lacking. From these four cases, some questions seemed to rise. Is the incidence of human listeriosis higher in a lower socio-economic group? Oakland has a large negro population, many of whom are of marginal or welfare status. If there be a higher incidence in poorer classes, why should this be? Could such groups have increased contact with animal hosts of infection? Could such groups have a lowered resistance to infection?

***ARCHIVES** introduces this "department" for its readers' views and observations. The editors will welcome reports or letters of pediatric and related-subject significance.*

The appearance of these four cases from similar age and racial and economic backgrounds poses an intriguing question. Are these of purest chance? Possibly so. In an effort to learn more about this, I contacted Dr. Paul D. Hoeprich regarding his personal views on the matter. Dr. Hoeprich recently wrote a splendid review article on human listeriosis.[4, 5] He commented he believed Seelinger's[6] original description of outbreaks of listeriosis in Germany did seem to involve a large proportion of children from lower economic brackets, but the point was not stressed. Dr. Hoeprich stated that most of his experiences were with indigent patients as seen in a teaching center, and hence would not necessarily reflect the true picture of incidence. However, this also raises a point: namely, that the bulk of reported cases in this country appear to come from county or charitable institutions, and teaching centers. Certainly, enough cases have been reported from lower economic groups to make one wonder. More information is sorely needed, and it is hoped that someone may be stimulated to search further in these areas.

In the four cases of listeria meningitis under discussion, it is regrettable that vaginal cultures were not obtained from the mothers. An attempt was made in one case, but the mother could not be located, which commonly happens in the case of indigent patients. It is felt by most authors that listeria infections of the newborn result from either intrauterine infection antepartum, or from vaginal sources at the time of birth. A further complicating factor is that if the mothers suffer from a generalized infection prior to birth, the symptoms may be minimal or absent.

Two additional cases of listerian meningitis are noted herein, with comments regarding the epidemiology, in the hope more light may be shed on the entire field of human listeriosis. I would be pleased to receive comments from any interested persons.

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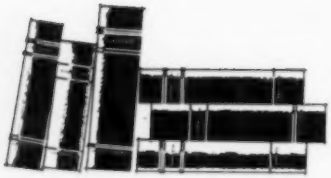

. . . Books

Edited by
MICHAEL A. BRESCIA, M.D.

RADIOGRAPHIC ATLAS OF SPECIAL SKELETAL DEVELOPMENT OF HAND AND WRIST, 2nd edition. William Falter Greulich and S. Idell Pyle. Stanford University Press. 1959. $15.

To the pediatrician the proper growth and development of their little patients is of paramount importance. Age, height and weight tables of which there are a great variety, are of aid but admittedly inadequate in covering all abnormal or disease conditions affecting growth.

As the authors point out, two factors in this country militate against a proper evaluation by tables (1) the heterogenicity of our national, and to some extent racial, origin and (2) the existence of early maturing and late maturing strains in our population, which influence the onset of puberty and the age at which preadolescent "spurt of growth" occurs.

This volume offers the pediatrician an aid in supplementing and perhaps supplanting table estimations, namely, X-ray studies of the hand and wrist at successive ages. It deals not only with normal standards, but with deviatons due to endocrine disturbances and illness. Male and female bone growth are considered seperately.

Although the average pediatrician has not received sufficient intensive training in the interpretation of X-ray to qualify him as an expert, this volume is so clearly written and beautifully and profusely illustrated with both film reproductions and diagrams that he should receive great assistance in the correct evaluation of growth and development. The information which it contains cannot be obtained in any other single volume in English. It is highly recommended. J. F. L.

THE NURSING OF MENTAL DEFECTIVES by CHARLES A. HALLAS. The William & Wilkins Company, Baltimore, Md.—Exclusive U. S. Agents. Cloth, pp. 182. Illustrated. Price $5.00.

The nursing of mental defectives is a growing science in our expanding interpretation of mental and emotional illness. Problems need definition in regard to the proper management of mental defective disorders. The author defines the subject very well.

This text has been meant for nurses but can readily serve allied workers in fields of education and social work.

This book considers mental deficiency fairly thoroughly. It includes the legal aspects, methods of examination, investigation, and education of the mental deficient. The book was written in England and there may be slight variations in our legal disciplines but the classification of mental disorders in relation to mental deficiency, methods of treatment, rehabilitation, and personality of the mental defective are standard. It is an excellent book for beginner's study in public health and rehabilitation. It is clear and easy reading.

I recommend this book to any person interested in mental defectives. ELSIE MECHTA, R.N.

SMALL PATIENTS. The Autobiography of a Children's Doctor. By ALTON GOLDBLOOM, M.D., Cloth. PP. 316. Price $4.50. J. B. Lippincott Co., Philadelphia and New York, 1959.

Dr. Goldbloom, a famous and well known Canadian pediatrician has written his life story. He is quite candid and realistic in telling his story which in no small measure increases the value of the book. His is a story which demonstrates how one can climb above all apparent obstacles both personal and environmental, if there is that innate ability and drive to succeed. His early life comprised an unstable home, uncertain economic status, inadequate education and vacillation between choice of careers. The story of his early life might better fit an individual in a reform school. Yet, once he set his sights on his future career there was no staying the man until he reached the pinnacle of his chosen field—the care of infants and children.

This book was of interest to me and I'm sure will be to medical historians for his intimate glimpses of some of the great names in pediatrics, such as Holt, Koplik, Schick, Schloss, etc. The author has a most subtle way of removing the cloak of glamour which surrounds these great names and shows that they too were human and subject to all the human foibles.

One gets the feeling on reading this autobiography that the subject is kindly, has done a great deal of good, is highly respected internationally and the picture of him that comes with the book shows him to be quite handsome, to boot. M. A. B.

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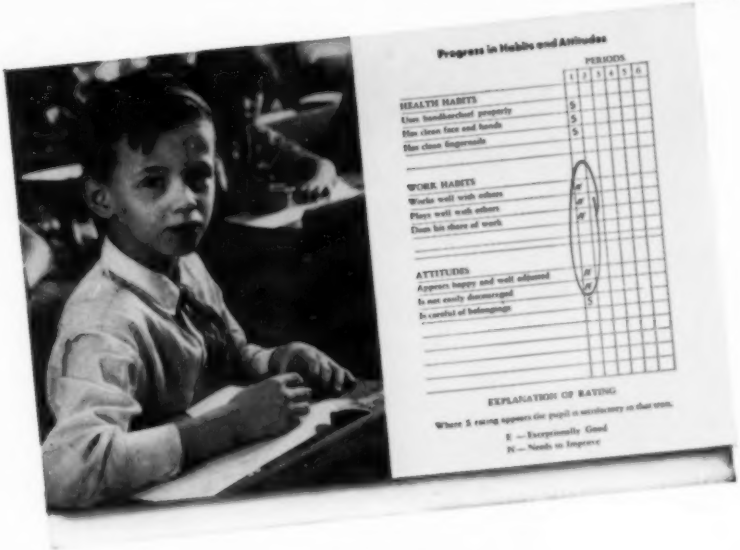

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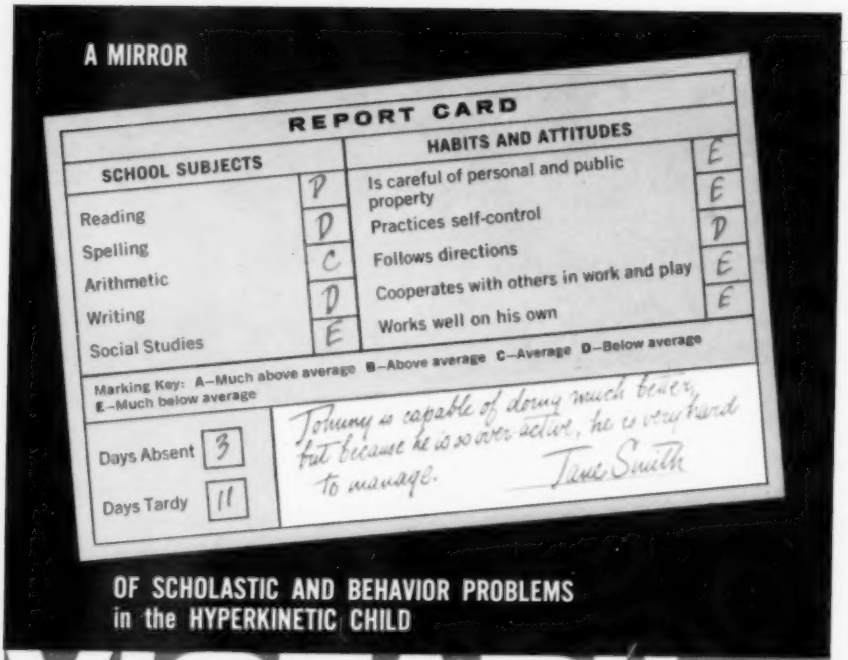
Division, Chas. Pfizer & Co., Inc.
Science for the World's Well-Being



An effective adjuvant in the treatment of hyperkinesis

DEPENDABLE PSYCHOTHERAPEUTIC POTENCY exerts a calming effect on the hyperactive, impulsive, destructive, aggressive, and difficult-to-manage traits of the hyperkinetic child.

"It is especially valuable in helping to calm the so-called 'hyperkinetic' child who seems to be on a constant merry-go-round."[1] *"This drug lessens tensions, relieves symptoms of disturbed autonomic function and calms hyperkinetic behavior."*[2]

FREEDOM FROM MENTAL STAGNATION especially significant in the school child.

"The relative freedom from side effects makes hydroxyzine quite valuable for anxious children and hyperkinetic children, especially those attending school, in whom drug-induced psychic impairment is undesirable."[3]

RECOMMENDED ORAL DOSAGE IN PEDIATRICS: under 6 years—50 mg. daily in divided doses; 6 years and over—50-100 mg. daily in divided doses.

SUPPLY: Vistaril Capsules—25 mg., 50 mg., 100 mg.; Vistaril Parenteral Solution—10 cc. vials and 2 cc. Steraject* Cartridges, each cc. containing 25 mg. hydroxyzine (as the HCl).

REFERENCES: 1. Eisenberg, B. C.: Clinical Medicine 5:897-904 (July) 1958. 2. Ayd, F. J., Jr.: Med. Arts and Sciences 2:37, 1957. 3. Ayd, F. J., Jr.: New York State J. 57:10 (May) 1957.

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